

A rare case of transposition of great arteries with an intact septum and aorto-pulmonary window

Sanjeev Singh^{1,4}, Arti Singh^{2,*}, Deigheidy Ehab Mahrous^{3,4}

¹Department of Anaesthesia and Intensive Care, School of Medical Sciences, College of Health Sciences

²KNUST Hospital, KNUST, Kumasi, West Africa

³Centre Chirurgicale Marie Lannelongue, Université Paris-Sud, France

⁴Department of Pediatric Cardiac Sciences, NHIMS, Bangalore, India

Email address:

artisingh_uk@yahoo.com (A. Singh)

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Abstract: Transposition of the great arteries with an intact ventricular septum and aortopulmonary window is an extremely rare anatomic combination associated with high morbidity and mortality. We report a case of a 24days neonate with dextro-transposition of the great arteries with intact ventricular septum and a large aortopulmonary window as a mechanism of inter-circulatory mixing.

Keywords: Aorto-Pulmonary Window, Arterial Switch Operation, Congenital Heart Defect, Transposition of the Great Arteries

1. Introduction

Dextro-looped transposition of the great arteries (D-TGA) accounts for 7 to 8% of all congenital cardiac malformation¹. About 25% of CHDs are considered critical congenital heart defects (CCHDs). Children with CCHDs need surgery or other procedures within the first year of life. CHD is the leading cause of birth defects, and accounts for more deaths in the first year of life than any other condition when infectious etiologies are excluded². Presentation and survival depends upon the admixture between systemic and pulmonary circulations. Conventional sites for shunting are atrial septal defect (ASD), patent foramen ovale (PFO), Ventricular septal defect (VSD) and Patent ductus arteriosus (PDA), rarely this could be an Aorto –Pulmonary Window (APW). Only few cases been reported with this association^{3,4,5}.

Aorto-pulmonary window leads to systemic pressure in pulmonary artery. Thus in TGA with intact IVS presence of non-restrictive APW prevents LV regression. At the same time, APW increases the risk of early pulmonary vascular disease if not operated early.

2. Case Report

A full term newborn with birth weight of 2.8 kg presented with complaints of rapid breathing, bluish

discoloration and poor feeding shortly after birth. Day 24th on clinical examination, baby was found to have tachypnea, retraction, tachycardia, hyper- active precordium, 3/6 ejection systolic murmur at left upper parasternal border and hepatomegaly. Echocardiographic examination showed situs solitus, levocardia, side by side great arteries, concordant atrio-ventricular connection, discordant ventricular-arterial connection, large secundum atrial septum defect, enlarged hypertensive right ventricle, large aortopulmonary window (type A), BD shunting coronaries from preferred facing sinus with good bi-ventricular function. Left ventricle was well preserved. After evaluation patient was operated.

3. Surgical Technique

Earlier reports have mentioned a modified technique of repair of APW along with ASO⁶. Midline sternotomy approach was selected. Exposure of the great vessels was undertaken and dissection of the pulmonary arteries was done. Cardiopulmonary bypass (CBP) was initiated with aortic and bicaval cannulation. Left atrial vent was inserted. Under ischemic arrest with cold anti grade cardioplegia the aorta and distal main pulmonary artery were transected. Aorta was transected at the level of AP window at about 8 mm above the level of coronary ostia. Coronaries were

implanted into the neo aorta. The MPA was reconstructed using an autologous pericardial patch and attached to the confluence of the pulmonary arteries. Branch pulmonary arteries were translocated anteriorly (Lacompt's maneuver). ASD was closed using autologous pericardial patch. Cross clamp was released, and assuring adequate coronary perfusions, sinus rhythm was achieved. Intra operative Trans Esophageal Echocardiogram showed good cardiac function and optimal repair. Total myocardial ischemic time and CPB time were 58 and 138 min respectively. Patient was transferred to the pediatric cardiac ICU in a good condition with minor inotropic support and mechanically ventilated.

4. Post-Operative Course

The patient had smooth post operative course. Child was extubated within 36 hours and shifted to ward on 3rd postoperative day (POD) without any adverse haemodynamic changes due to extubation keeping in mind⁷. Repeated echo after surgery revealed no ventricular out flow tract obstruction and normal cardiac function. Ward stay was uneventful and patient was discharged on 11th POD on furosemide, enalapril and sildenafil.

5. Discussion

Aorto-pulmonary window is a rare cardiac defect. Abnormal separation of the aortic sac by the aortic-pulmonary septum during embryologic development is thought to be the cause of APW⁸. The location limited between the two semilunar valves and the branch pulmonary arteries of varying size. Various morphological forms led to the development of several classifications of APW. Richardson's classification may be used as a guide for the appropriate surgical approach. 11 cases of APW associated with TGA were reported in the literature to our knowledge. The risk of surgery in this group is substantial with 6/11 cases survived. All of them had arterial switch operation between age of 7 days and 3 years.

Surgical correction of complete TGA with intact inter ventricular septum comprises of Arterial Switch Operation (ASO) & Atrial Switch (Senning's or Mustard). ASO is surgery of choice as it re-establishes ventriculo-Arterial concordance. But it has to be performed in the first 4 weeks, prior to unfavorable changes in left ventricle (LV). For late presenting babies or with coronary anatomy making ASO difficult, atrial switch is considered, with acceptable short-term morbidity and mortality. Arterial switch surgery seems to be the best surgical option for this complex cardiac lesion. Co-existing APW provided for enough mixing and pressure load on LV. APW leads to accelerated progression of pulmonary vascular disease as the flow to pulmonary circulation is un-restricted. Early recognition of

transposition of the great arteries with aorto –Pulmonary window gave us an opportunity to mend the defect.

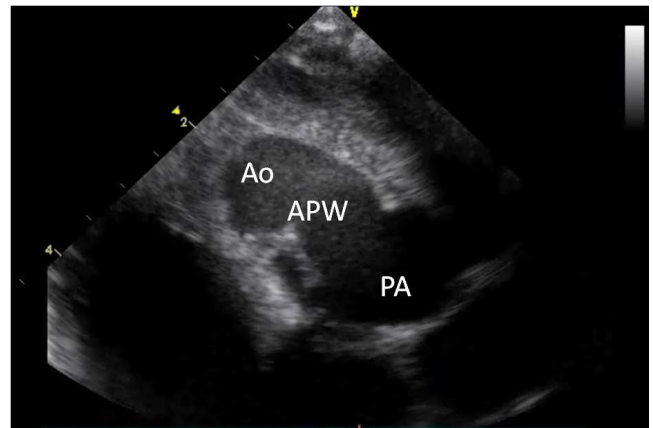


Figure 1. Suprasternal 2D Echocardiography View showing a large communication (APW) aortopulmonary window between ascending aorta (Ao) and pulmonary artery (PA).

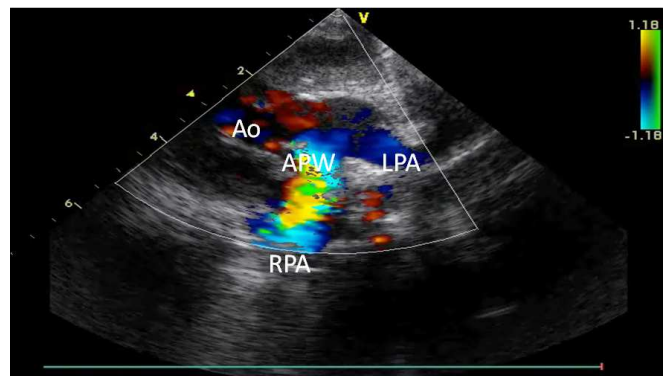


Figure 2. High Parasternal Short Axis View with Color Doppler showing unrestricted flow from aorta (Ao) to pulmonary artery (PA). Ao. Aortopulmonary window (APW).

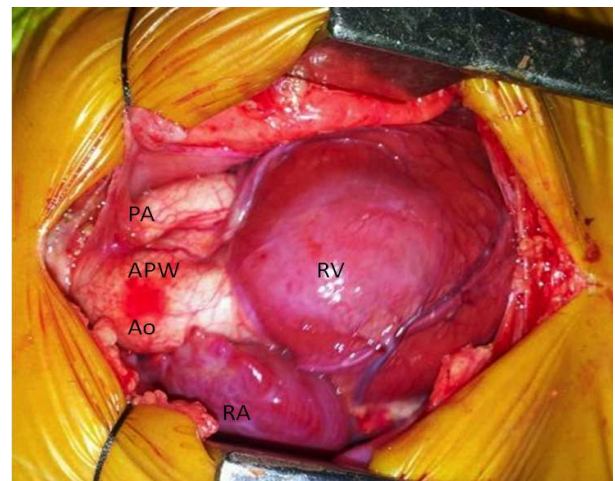


Figure 3. Surgeon's view showing right atrium (RA), right ventricle (RV), Aorta (Ao) from right ventricle (RV), Communication between Aorta and pulmonary artery (PA) anteriorly aorto-pulmonary window (APW).

Table 1. Out-come of the 12 reported cases of TGA with APW.

No.	Author	Year of surgery	Age at surgery	Diagnosis	Surgery	Out-come
1	Tiraboschi et al.	1988	7 mo	TGA, double APW, VSD	-	Death
2	Krishnan et al.	1991	3 mo	TGA APW VSD	Senning repair	Death
3	Tirado et al.	1993	7 d	TGA VSD APW	Direct suture, PAB	Death
4	McElhinney et al.	1995	8 d	TGA ASD APW	ASO	Death
5	Marangi et al.	1996	7 d	TGA APW PDA	ASO	Alive
6	Backer et al.	1996	3 yr	TGA APW	ASO	Alive
7	Duca et al.	2002	26 d	TGA APW	ASO	Alive
8	Marwah et al.	2005	11 mo	TGA APW	ASO	Alive
9	Adluri et al.	2005	8 d	TGA APW	ASO	Alive
10	Das et al.	2006		ccTGA APW	-	Death
11	Najm et al	2008	32 d	Dextrocardai, TGA, APW, PDA	ASO	Alive
12	Singh et al. (current case)	2013	26 d	D-TGA, APW, ASD	ASO with ASD closure	Alive

TGA = transposition of great arteries, APW = aortopulmonary window, ASD = atrial septal defect, PAB = pulmonary artery band, ASO = arterial switch, ccTGA = congenitally corrected
 (-)= data not available/ not operated .

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